MÆDICA - a Journal of Clinical Medicine

STATE OF THE ART

MAEDICA – a Journal of Clinical Medicine
2016; 11(4):316-319

Prenatal Diagnosis in Great Artery Trasposition and Implications in Postnatal Outcome

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ABSTRACT

Great artery transposition, one of the most frequent and severe cyanotic heart malformations, represented the subject of many studies and research up to this moment. Although postnatal period is critical in this pathology, with correct management patients can benefit from complete surgical correction - arterial switch operation with good long term evolution. Prenatal diagnosis of Great Artery Transposition has an important contribution in choosing the appropriate treatment at the right time in postnatal period so that specific complications resulting from delaying the initiation of specific therapies can be avoided. This article proposes a review of existing data at this moment related to the importance of prenatal diagnosis in Great Artery Transposition and underlines how an accurate fetal diagnosis influences the decision of establishing the appropriate treatment in postnatal life for the children with this type of malformation and the complications that can be avoided.

Keywords: Prenatal diagnosis in great artery transposition

INTRODUCTION

Transposition of the great vessels (TGA) is one of the most common cyanotic heart malformations, representing 5% to 7% of all cardiac malformations that result in death without proper treatment of approximately 30% of cases in the first week of life (1).

In this anomaly, the aorta arises from the morphological right ventricle (RV), and the pulmonary artery arises from the morphological left ventricle (LV), resulting in ventriculoarterial discordance. Complete transposition of the great arteries is also known as d-TGA; the "d-" refers to the dextroposition of the bulboventricular loop, the position of the RV, which is on the right side. The aorta also tends to be on the right and anterior, and the great arteries are parallel rather than crossing as they do in the normal heart. Because the systemic and pulmonary circulations run in parallel, there has to be a communication between the two, either with an atrial septal defect, a ventricular septal defect (VSD), or at the great

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Article received on the 12rd of December 2016. Article accepted on the 22rd of December 2016.

arterial level - patent ductus arteriosus (PDA), to support life. These connections allow systemic blood to enter the pulmonary circulation for oxygenation and allow oxygenated blood from the pulmonary circuit to enter the systemic circulation. The most common associated lesions are VSD, which occurs in almost half of the cases, pulmonary outflow tract obstruction, and, less commonly, coarctation of the aorta (2). In approximately one third of patients with transposition of the great arteries, the coronary artery anatomy is abnormal, with a left circumflex coronary arising from the right coronary artery (22%), a single right coronary artery (9.5%), a single left coronary artery (3%), or inverted origin of the coronary arteries (3%) representing the most common variants (3).

The initial management consists of stabilizing the heart and lung function to ensure the mixing between the pulmonary and systemic circulation by keeping the ductus arteriosus open with prostaglandin E1 infusion and/or atrial balloon septostomy (BAS) (4).

Newborns with TGA have systemic and pulmonary circulation in parallel; thus, mixing of both circulations is essential to maintain appropriate oxygen saturation level. The shunt flow through a PDA, VSD, and ASD may result in hemodynamic instability despite of the decrease of the pulmonary arterial resistance. It is considered that using an ASD to provide the appropriate oxygen saturation is better and hemodynamically more stable than prostaglandin infusion for keeping the PDA open.(5)

FIGURE 1. Fetal echocardiographic image of patient with transposition of the great arteries

It is generally believed that infants with TGA should undergo arterial switch operation (ASO) in the first two weeks after birth. Nevvazhay *et al.* proposed that ASO should be carried out as early as possible on infants with TGA, without performing baloon atrial septostomy (BAS). Anderson *et al.* suggested that ASO performed on the 3rd day after birth could avoid the deterioration of left ventricular function, reduces postoperative complications and mortality, and thus effectively reduces hospital stay and costs. An alternative operation was subsequently developed by Mustard, who excised the atrial septum and used synthetic material to create the baffle. Sometimes an atrial switch is preceded by either the Rashkind balloon atrial septostomy or surgical atrial septectomy (Blalock-Hanlon). Both atrial switch procedures provide excellent midterm clinical results but in the long term are associated with important sequelae. At present, the specific timing of surgery remains controversial and depends on the type of shunt that provide the systemico-pulmonary mixing. Atrial septostomy should be reserved for the cases that cannot be operated in the first days of life, thus prolonging the time before surgery to approximatively 1 month. The most common complications of this procedure are: vascular lesions, arrhythmias, atrial perforation, cardiac tamponade and an increased risk of cerebral embolism (6).

Despite the progresses that have been made in fetal ultrasound, cardiac malformations are diagnosed in approximatively 50% of cases, TGA with intact ventricular septum having a rate of 50-70% prenatal diagnosis in Europe (7).

Prenatal diagnosis of transposition of the great arteries is associated with a significant reduction in both preoperative and postoperative mortality, a decrease in the rate of metabolic acidosis and multiorgan failure during the neonatal period, reduced need for ventilatory support, and shorter hospitalization time (8).

In a study that was conducted over 10 years (1987-1998) in Paris, which included 68 newborns with prenatal diagnosis of TGA and 250 newborns with postnatal diagnosis of TGA was observed that the transfer in pediatric cardiology clinic was significantly earlier in the fetal diagnosed group (2.2 hours *versus* 73 hours); mechanical ventilation was required more frequently in the neonatal diagnosed group (38%) *versus* the fetal diagnosed group (17.6%); metabolic

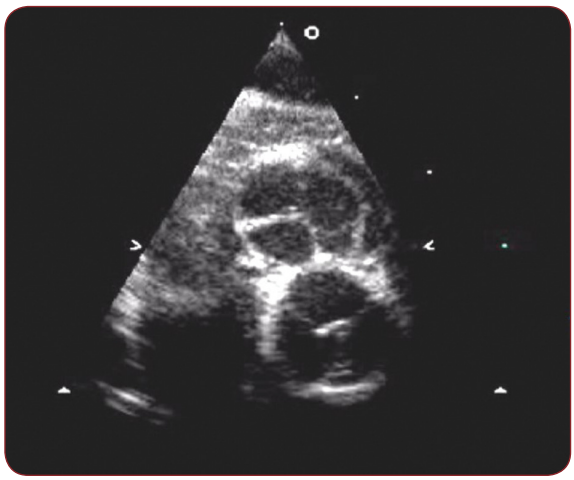

FIGURE 3. Transthoracic echocardiography - parasternal short-axis view demonstrating the commissural malalignment between the commissures of the aortic valve and pulmonary valve

acidosis (pH <7.10), multiorgan failure and neurological impairment were significantly more frequent in neonatal group (p = 0.02). Postoperative mortality was significantly higher in the neonatal group 8.5% vs. 0 in prenatal group and total hospital stay was longer in the neonatal group 30±17 days *versus* 24 ± 11days (9).

Another study conducted over a period of 20 years (1992-2011) at Boston Children's Hospital including 340 infants with a diagnosis of TGA without other associated malformations of which 81 (24%) patients diagnosed prenatally, showed that the rate of prenatal diagnosis increased during the study from 6% to 41%; patients diagnosed prenatally benefited earlier of atrial balloon septostomy and fewer required mechanical ventilation. Postoperative mortality rate was not significantly different between the group diagnosed prenatally *versus* the group diagnosed postnatally but there were significant differences in preoperative period, atrial septostomy was made earlier in prenatal group and the need for mechanical ventilation was lower in the same group (7).

Postoperative neurological complications of patients with TGA were the subject of a retrospective study on 170 patients from which 53 (31%) having prenatal diagnosis in 2005-2011. There were observed episodes of preoperative seizures and stroke, seizures were diagnosed clinically and the stroke by MRI or CT. Seizures have been identified in 10 of the patients diagnosed postnatally *versus* 0 of those with prenatal diagnosis and in the postnatal group stroke was identified in 9 patients *versus* 0 in the prenatal group. It thus concluded that the association between neurological complications and the time of diagnosis is significant for patients with TGA (10).

Prenatal risk factors for poor outcome in neonates were followed in a study on 50 fetuses diagnosed with TGA at the University of Alberta and University of Minnesota between 2003 and 2016. It has been found that: a reversed diastolic flow on PDA, a hypermobile redundant interatrial septum in fossa ovalis area, a restrictive fossa ovalis of ≤6 mm predicted urgent need for BAS within 2 hours after birth. Ratio of fossa ovalis size to interatrial septum of ≤0.3 predicted urgent need for BAS within 2 hours after birth. 22 of 31 (71%) that required BAS had thick atrial septum. 19 (100%) that did not require BAS had thin atrial septum. In conclusion, effective orifice of fossa ovalis and ratio of fossa ovalis to interatrial septum are predictors for the need for urgent BAS postnatally. Increased thickness of atrial septum causes restriction of effective orifice of the fossa ovalis necessitating urgent BAS postnatally (11).

Also, fetal ultrasound can detect associated cardiac malformations with this aspect in determining the best therapeutic implications both in immediate postnatal period and in advising parents, providing important data on the probable prognosis of the child on long-term, but the accuracy of the diagnosis depends largely on the experience of the examiner. Based on the data from fetal echocardiography the parents can be advised on the prognosis of the child and what they should expect after birth.

It is mentioned that almost 50% of infants with TVM associate other cardiac abnormalities including coronary artery anomaly which makes de surgical intervention more difficult. VSD is present in 40-45% of cases of TGA; detecting a fetal ultrasound performed at large DSV near birth practically exclude the need for immediate postnatal atrial septostomy. It should be noted that based on fetal ultrasound can appreciate quite correct the optimal surgical time for correction: in the case of a large VSD intervention (arterial switch) may be delayed up to two months, while small shunts at ASD / VSD require early correction at the age of 7-10 days. In some cases TGA is associated with large VSD and outflow

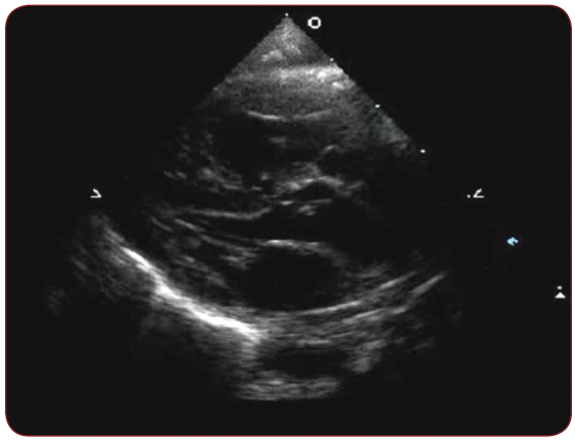

FIGURE 2. Transthoracic echocardiography, parasternal long-axis view with the typical parallel orientation of the great valves

tract obstruction in LV and corrective surgery may be performed around the age of 6 months.❑

CONCLUSION

Prenatal diagnosis is very important in TGA because of its multiple implications on the establishment of appropriate therapy in the postnatal period, significantly influencing the type and number of postnatal complications.

Given the severity of this malformation and the need to establish an early adequate therapy, fetal ultrasound provides important information on the hemodynamic expected status in postnatal period, the need of emergency septostomy, associated anomalies, significantly helping to manage these complex cases.

Echocardiographic parameters followed in prenatal period may bring important clues on cases that will require invasive procedures without assigning an absolute value to prenatal echocardiography. After initiating the infusion of prostaglandin, subsequent treatment decisions are made based on data provided by the postnatal echocardiography in tight conjunction with clinical data.❑

Conflict of interests: none declared.
Financial support: none declared.

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